Original Article

www.ijrhs.com
ISSN (o):2321–7251

Immune thrombocytopenic purpura: A 5-year study, our experience at a tertiary care centre

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Submission Date: 09-01-2014, Acceptance Date: 14-01-2014, Publication Date: 31-01-2014

How to cite this article:

Vancouver/ICMJE Style

Neelaveni N, Jeshtadi A, Sri S, ML, Prathima P. Immune thrombocytopenic purpura: A 5-year study, our experience at a tertiary care centre. Int J Res Health Sci [Internet]. 2014 Jan31;2(1):316-9. Available from http://www.ijrhs.com/issues.php?val=Volume2&iss=Issue1

Harvard style

Neelaveni, N., Jeshtadi, A., Sri, S., M.L., Prathima, P. (2014) Immune thrombocytopenic purpura: A 5-year study, our experience at a tertiary care centre. *Int J Res Health Sci*. [Online] 2(1). p. 316-9. Available from: http://www.ijrhs.com/issues.php?val=Volume2&iss=Issue1

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Abstract:

Introduction: The disease that was formerly termed idiopathic thrombocytopenia (ITP) is now called immune thrombocytopenia. Immune mediated thrombocytopenia is caused by autoantibodies or alloantibodies. Auto- antibody-related immune thrombocytopenic purpura (ITP) is the most common cause of immune thrombocytopenia. It can be acute or chronic, idiopathic (primary) or secondary. Acute ITP occurs mostly in children and is commonly associated with viral infections, while adult ITP may be primary or secondary to infections, other systemic diseases or drugs and is a diagnosis of exclusion. Patients often present with petechiae, bruising, mucosal bleeding and rare presention of thromboembolic episodes. **Objective:** This is a 5-year retrospective study with analysis of ITP cases, age and sex incidence, clinical and hematological findings done at a tertiary care centre. **Results**: Total number of cases was 38. Age group ranged from 11-40 years with mean age of 26.28 years. Male to female ratio was 1:2.8.Epistaxis was the commonest clinical presentation. Hematological data showed platelet count below 1.5 lakhs per decilitre of blood with normal and giant platelets in morphology. Bone marrow examination showed normal and increased number of megakaryocytes.

Key words: Autoantibodies; Bone Marrow Examination; Hemorrhagic Disorder; Immune Thrombocytopenic Purpura; Splenectomy

Introduction

Once regarded as idiopathic, immune thrombocytopenia (ITP) is now understood to have a complex pathogenesis, involving the evolution of antibodies against multiple platelet antigens leading to reduced platelet survival as well as impaired platelet production [1].

Both acute and chronic forms of disease can be distinguished. In children acute ITP is often associated

with a viral or bacterial infection and generally resolves spontaneously within 6 weeks. Approximately 20% of children with acute ITP progress to the chronic form. In contrast ITP in adults is generally chronic and often requires treatment [2].

Immune thrombocytopenic purpura (ITP) patients often present with petechiae, bruising, and mucosal bleeding. A life-threatening hemorrhage is rare, especially in acute ITP. Immune thrombocytopenic purpura may be manifested in acute or chronic forms and in primary or secondary forms. A wide variety of therapeutic regimens are currently used for the treatment of ITP due to autoantibodies [3].

The main diagnostic procedure for ITP is based on patient history, physical examination, complete blood count, and peripheral smear examination. Peripheral blood examination reveal normal-to-large platelets with normal red cell and white cell morphologies.Bone marrow aspirates show normal,or increased number of megakaryocytes [3].

Immune thrombocytopenic purpura is a diagnosis of exclusion. Other causes of thrombocytopenia should be ruled out, and these include drug-induced thrombocytopenia, bone marrow failure, inherited thrombocytopenia, microangiopathic and leukemic thrombocytopenias, and other disorders associated with thrombocytopenia [4].

Materials and Methods

This was a 5-year study done at a tertiary care centre from January 2009 to December 2013.A total of 13508 hemograms were done of which 1223(9.03%) cases with low platelet count (less than 1.5 lakhs per dl of blood) were diagnosed. Clinical data, hematological and bone marrow aspiration findings were evaluated. Bone marrow examination was done for 160(13.08%) cases. After exclusion of other diseases, 38 cases of immune thrombocytopenic purpura were confirmed.

Results

In this retrospective study we found that ITP was more common in females with a male to female ratio being 1:2.8.Most of the cases were young, in the age group of 21-30 years.

General examination of the patients revealed splenomegaly in 18(47.36%) cases, hepatomegaly in 4(10.52%) cases and cervical lymphadenopathy in 2(5.26%).

Table-1: Age and sex distribution of cases

Age group	No. of females	No. of males	Total number of cases
0-10	0	0	0
11-20	6(15.78%)	2(5.26%)	8(21.05%)
21-30	13(34.21%)	5(13.15%)	18(47.36%)
31-40	9(23.68%)	3(7.89%)	12(31.57%)
41-50	0	0	0
Total number of cases	28(73.68%)	10(26.31%)	38

Clinical Presentation:

The most common clinical presentation was epistaxis. We also noticed that there were substantial number of cases presenting with menorrhagia.

Table 2: Clinical Presentation

Presenting feature	No. of cases	Percentage
1. Epistaxis	22	57.89%
2. Menorrhagia	6	15.78%
3. Purpura & Petechial rash	4	10.52%
4. Melena	3	7.89%
5. Bleeding gums	2	5.26%
6. Fever & Hematuria	1	2.63%

Table 3: Platelet Count

Platelet count	**No. of cases**
<10000	02(5.26%)
10000-20000	08(21.05%)
21000-30000	11(28.94%)
31000-40000	04(10.52%)
41000-50000	03(7.89%)
51000-60000	04(10.52%)
61000-70000	03(7.89%)
71000-80000	02(5.26%)
81000-90000	01(2.63%)
91000-100000	00
Total number of cases	**38**

Table 4: Peripheral smear

Cells	**Morphology**	
Red Blood Cells	Normocytic normochromic (47.36%)	Microcytic hypochromic 20(52.63%)
White Blood Cells	Normal 38(100%)	
Platelets	Normal 26(68.42%)	Giant Platelets 12(31.57%)

Hematological analysis showed normocytic normochromic blood picture in 18 cases (47.36%) and microcytic hypochromic blood picture in 20 cases (52.63%). White blood cells showed normal morphology and counts in 38 cases (100%). Platelets showed normal size in 26 cases (68.42%) and giant platelets in 12 cases (31.57%).

Table 5: Bone marrow Examination

Cell Lineage	**Morphology**	
Erythroid	Normoblastic maturation 20(52.63%)	Micronormoblastic 18(47.36%)
Granulocytic	Orderly and normal maturation 38(100%)	
Megakaryocytic	Normal 27(71.05%)	Increased 11(28.94%)

Bone marrow examination showed normoblastic maturation in 20 cases (52.63%) and micronormoblastic maturation in 18 cases (47.36%). Granulocytic cell lineage showed orderly and normal maturation in 38 cases (100%). Megakaryocytic cell lineage showed normal number in 27 cases (71.05%) and increased number of megakaryocytes in 11 cases (28.94%).

Discussion

Idiopathic thrombocytopenic purpura (ITP, also first known as primary immune thrombocytopenic purpura) is a hematologic disorder for which appropriate diagnostic and treatment strategies are uncertain [5].

Idiopathic thrombocytopenic purpura (ITP) is an acquired hemorrhagic disorder characterized by thrombocytopenia that is defined as a platelet count less than 150 × 109/L (150,000/mcL), a purpuric rash, normal bone marrow, and the absence of signs of other identifiable causes of thrombocytopenia. ITP is classified as acute or chronic, with the latter defined as the persistence of thrombocytopenia for more than 6 months from the initial presentation of signs and symptoms.

Immune thrombocytopenic purpura (ITP) is an autoimmune disorder in which, for reasons that remain unclear, platelet surface proteins become antigenic and stimulate the immune system to produce autoantibodies and cytotoxic T lymphocytes. This results in immune-induced platelet destruction and suppression of platelet production. What causes the loss of tolerance to one's own platelets remains unclear and is likely to be a result of a number of different cooperating factors including genetics (polymorphism in selected genes) and environment events (virus and bacteria-associated ITP) [2].

ITP is estimated to be one of the most common acquired bleeding disorders encountered by pediatricians, with the incidence of symptomatic disease being approximately 3 to 8 per 100,000 children per year. Acute ITP is more prevalent among children younger than 10 years of age, affects males and females equally, and is more prevalent during the late winter and spring. Chronic ITP affects adolescents more often than younger children, with females being affected more frequently than males. Unlike acute ITP, it does not show a seasonal predilection. Patients who have chronic ITP are more likely to exhibit an underlying autoimmune disorder, with up to one third having clinical and laboratory manifestations of collagen-vascular disease [6].

Treatment is generally required for chronic ITP or for symptomatic patients with low platelet counts. Initial treatments include corticosteroids, intravenous immunoglobulin (IVIG), or anti-D immunoglobulin when indicated. Splenectomy is effective in most patients who are not responding to corticosteroids or IVIG. Immunosuppressive drugs and anti-CD20+ have been used successfully in treating patients with refractory ITP. Thrombopoietic drugs are the new promise in stimulating platelet production [7].

Ours is a 5-year retrospective study done at a tertiary care centre. A total of 13508 hemograms were done of which 1223(9.03%) cases with low platelet count (less than 1.5 lakh per dl of blood) were diagnosed.Bone marrow examination was done for 160(13.08%) cases. After evaluation of patient's history, physical examination, complete blood count, peripheral smear examination and bone marrow examination and

exclusion of other diseases, 38(3.10%) cases of immune thrombocytopenic purpura were confirmed.

In our study age group ranged between 11-40 years with mean age 26.28 years with female predominance and male to female ratio 1:2.8. The most common clinical presentation was epistaxis. We also noticed that there were substantial number of cases presenting with menorrhagia. Purpura, petechial rash, melena, bleeding gums, fever and hematuria were other presenting features. General examination revealed splenomegaly in 18(47.36%) cases, hepatomegaly in 4(10.52%) cases and lymphadenopathy in 2(5.26%) cases and no significant findings in 14(36.84%) cases. In our study criteria for thrombocytopenia was platelet count below 1.5 lakhs cells per dl of blood.

Platelet count <10000 in 2 cases (5.26%),10000-20000 in 8 cases (21.05%), 21000-30000 in 11 cases (28.94%), 31000-40000 in 4 cases (10.52%), 41000-50000 in 3 cases (7.89%), 51000-60000 in 4 cases (10.52%), 61000-70000 in 3 cases (7.89%),71000-80000 in 2 cases (5.26%), 81000-90000 in 1 case(2.63%). Platelets showed normal size in 26 cases (68.42%) and giant platelets in 12 cases (31.57%).

Bone marrow examination of cases showed normal number of megakaryocytes in 27 cases (71.05%) and increased number of megakaryocytes in 11 cases (28.94%).

Conclusion

Immune thrombocytopenic purpura is an uncommon condition that tends to resolve spontaneously within 6 months of presentation. Although usually self-limited, ITP can be a worrisome diagnosis for families and clinicians. Serious hemorrhagic complications of ITP seldom occur, regardless of whether symptoms persist beyond 6 months. Our results suggest that cases of ITP show varied clinical differences in bleeding symptoms, previous acute illness, age, and presenting platelet count. In practice, physicians may need to consider these characteristics when advising their patients and families about potential resolution and management of ITP.

Source of Funding: Nil

Source of Conflict: Nil

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